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TELANGIECTATIC OSTEOSARCOMA-AN IMAGING SPECTRUM

Radiology

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ABSTRACT

Context: Telengiectatic osteosarcomas (TOS) account for 2% to 12% of osteosarcomas. They have an osseous origin in the medulla of metaphysis of long bones; distal femur being most common site. In majority of tumor volume consists of large haemorrhagic or necrotic cavities accounting for 90% of lesion and small proportion of solid tissue. The walls, septae around haemorrhagic spaces are thick and nodular. Due to paucity of literature this study was undertaken.

Aim: This study is aimed at delineating the imaging features of Talengiectatic osteosarcomas

Methods And Material: All histopathologically proven cases of osteosarcoma attending our hospital were analysed. Six of the 35 cases were of TOS sub-type. Their imaging features- the bone-matrix consistency, zone of transition, fluid-fluid-level, periosteal reaction, soft-tissue and sclerosis were specifically studied.

Results: All 6 were males and most were in age-group of 10-20 years. Three tumours were in femur and one each in humerus, tibia and radius. All were situated in metaphysis of long bones. Lesions were predominantly lytic with expansion of bone with cortical destruction soft tissue extension. There was periosteal reaction in the form of interrupted lamellar, sunburst, Codman's triangle. 50% cases showed faint focal sclerosis at periphery. FFL were small and non-uniform with irregular walls and were observed in all cases.

Conclusions: Telengiectatic osteosarcoma is a rare variety of Osteosarcomas. It has to be differentiated ABC. Lytic lesion, wide zone of transition and malignant periosteal reaction is usually observed in TOS. FFLs are usually small with irregular septae. Presence of sclerosis is diagnostic of TOS.

KEYWORDS

INTRODUCTION:

Osteosarcoma is the second most common malignant bone tumor occurring mostly in 10-25 year- age group. Though knee is the most common site, it can also occur in other long bones. Telangiectatic osteosarcomas (TOS) account for 2% to 12% of osteosarcomas[1]. TOS was first described by Pagets in 1854. Gaylord used the term 'Malignant bone aneurysm' to refer to the same condition in 1903[2]. They have an osseous origin in the medulla of metaphysis of long bones; distal femur being most common site. In TOD, majority of tumor volume consists of large haemorrhagic or necrotic cavities accounting for 90% of lesion and small proportion of solid tissue. The walls, septae around haemorrhagic spaces are thick and nodular containing malignant cells that produce osteoid. Prognosis of TOS is worse than conventional osteosarcoma. In recent years due to advancement of treatment modalities 5-year survival rate improved to 67%. Due to paucity of literature the imaging features of TOS are analysed and the literature is reviewed in this study.

MATERIAL AND METHODS

35 consecutive cases of Osteosarcomas were studied for their imaging features. Out of these, TOS accounted for 17%. The bone-matrix consistency, zone of transition, fluid-fluid-level, periosteal reaction, soft-tissue and sclerosis are specifically studied on all available imaging modalities.

OBSERVATIONS

In present study Osteosarcoma was most prevalent bone tumor contributing to 70% (35 cases) of the total cases. Osteoblastic osteosarcoma was the most common subtype contributing 22 cases (62.5%) followed by Telangiectatic osteosarcoma contributing to 6 cases (17.1%) [Table-1].

Table-1: Sub Types Of Osteosarcomas.

TYPE	NO. OF CASES	PERCENTAGE
OSTEOBLASTIC	22	62.8%
CHONDROBLASTIC	2	5.7%
TELANGIECTATIC	6	17.1%
PARAOSTEAL	4	11.4%
PERIOSTEAL	1	2.8%

All 6 were males and most were in age-group of 10-20 years. Three tumours were in femur and one each in humerus, tibia and radius. All were situated in metaphysis of long bones. Lesions were predominantly lytic with expansion of bone with cortical destruction soft tissue extension. There was periosteal reaction in the form of interrupted lamellar, sunburst, Codman's triangle .50% cases showed faint focal sclerosis at periphery. FFL were small and non-uniform with irregular walls and were observed in all cases.

Table-2: Imaging Features Of The 6 Cases Of TOS

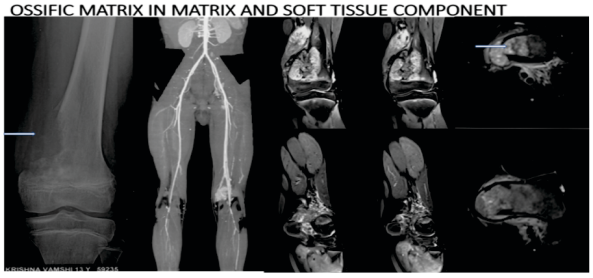
case	site	Lytic/ sclerotic/ mixed	Zone of transition	FFL	PR	Soft Tissue	Sclerosis
1	Metaphysis tibia	mixed	wide	Small, Non-uniform	+	+	+
2	Metaphysis radius	lytic	wide	Small Non-uniform		+	-
3	Metaphysis, femur	lytic	narrow	Large, Non-uniform	+	+	-
4	Metaphysis, femur	lytic	wide	Large, non-uniform	+	+	faint
5	Metaphysis, femur	lytic	wide	Multiple, small, non-uniform	+	+	+
6	Metaphysis, femur	lytic	wide	Large, non-uniform	+	+	-

DISCUSSION

Talengiectatic osteosarcomas occur in adolescents and young adults (3 years -67years) with mean of 20 years and shows male preponderance. All our patients were males.These TOS originate from metaphysis of long bones. The distal femur constituted 48% of

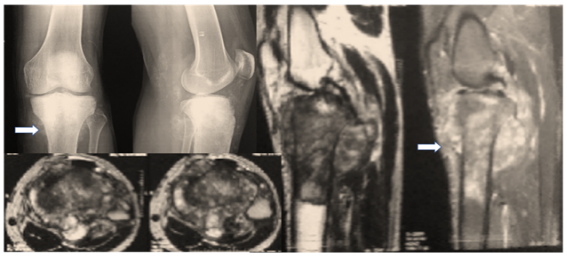
cases. Proximal humerus, proximal tibia are other sites in our series [Fig-1]. Usually geographic bone destruction with wide zone of transition and endosteal scalloping is observed. But it may be moth-eaten; permeative with zone of transition may be narrow, sclerotic/non-sclerotic. Out of our 6 patients, one case had narrow zone of transition. Aneurysmal expansion can occur which may be mild, moderate or marked. Cortical destruction and soft tissue component may be appreciated. Osteoid matrix mineralization is seen in 58% of cases. It is subtle and not appreciated on radiograph. Mostly the mineralization is seen at periphery or in trabeculae which contain tumor cells. Osteoid is usually not appreciated on radiograph and CT may be helpful to identify it as faint lace like tissue. We had similar observation. 50% cases had sclerosis at periphery. The osteoid and Chondroid mineralization may be observed in soft tissue component [Fig-1].

FIG 1 13M ECCENTRIC,EXPANSILE GEOGRAPHIC LYTIC LESION WITH WIDE ZONE OF TRANSITION.FFLS ARE SEEN.CLOUD LIKE OSSIFIC MATRIX IN MATRIX AND SOFT TISSUE COMPONENT



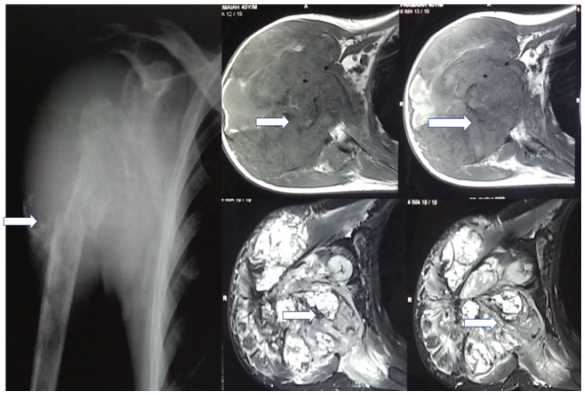
The cystic nature of tumor is reflected by lower attenuation at the centre as compared to periphery at CT. Often it is lower than the muscle. On MRI then tumor appear bright on T2W images. The haemorrhagic component is appreciated well on MR. The fluid- fluid levels and soft tissue component are appreciated both on CT and MRI. All our cases have FFL and soft tissue component [Fig-2].

FIG 2 19M CORTICAL DESTRUCTION,PERIOSTEAL REACTION(ARROW) ,SOFT TISSUE COMPONENT



On nuclear scan the tumor gives *Donut sign* that is central photogenic area and peripheral increased. The margin of TOS is mostly infiltrative or non- encapsulated [Fig 3]. 5 out of 6 had diffuse soft tissue. Protrusion is mostly focal in contrast to ABC where there is expansion remodelling of bone is more than 33% of circumference, [note focal protrusion in fig 3]. An intact cortex and associated fibrosis produce hypodense and hypointense rim surrounding the ABC[3].

FIG 3 40M OSSIFIC MATRIX IS SHOWN AS ARROW.FFLS ARE UNUNIFORM AND LARGEST FFL IS SMALL



In few cases there may be capsule. A retrospective analysis of 40 cases of TOS, they found infiltrative margin in 48%, defined margin with no capsule in 41% and defined with pseudocapsule in 11%. Joint involvement was seen in 44% cases[1] [Fig-4&5].

FIG 4 19M FFLS.THIN AND THICK SEPTAE AND PSEUDOCAPSULE(ARROW)

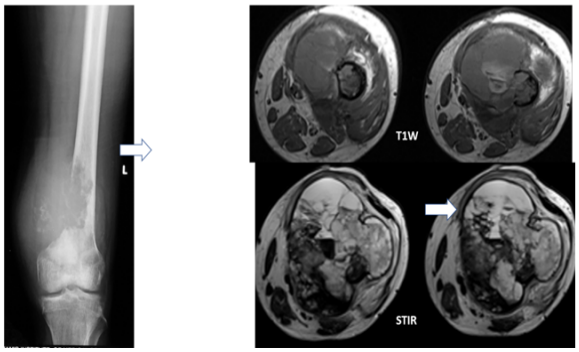
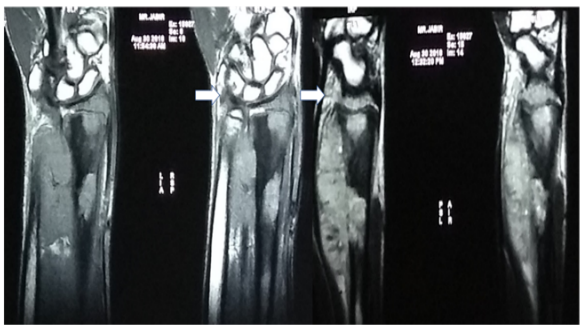


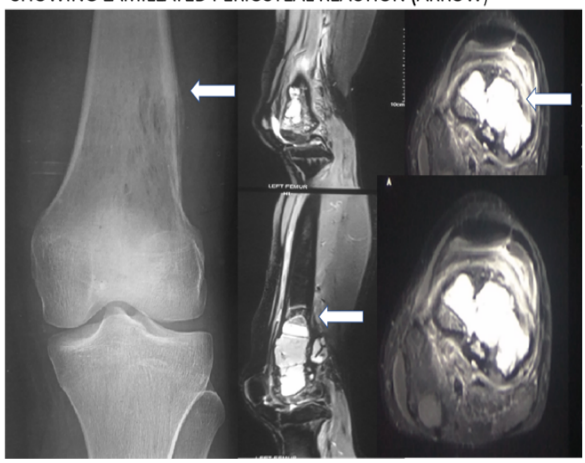
FIG 5 17M T1W,T2W IMAGES SHOWING LYTIC LESION IN RADIUS WITH LARGE SOFT TISSUE COMPONENT. EXTENSION TO WRIST JOINT IN SAGITTAL T1 AND T2W IMAGES



Ratio of FFL to tumor occupancy is least in malignant lesion as compared to primary or secondary ABC. In a series of 83 patients with bone lesion containing FFLS, it was found that proportion of lesion fluid level was inversely related to likelihood of malignancy[4]. Hong Yu et al showed in their study of 47 patients that the maximum length of the largest FFL to maximum length of tumor in sagittal plane was higher in benign than malignant tumor. They suggested that when the ratio is more than 41.5% the masses were considered benign with sensitivity of 73% and specificity of 85%[5]. All our cases showed FFL [Fig-3]. These FFL are small and of varying sizes. 50% cases had one or two large FFL, rest were small in size and non-uniform.

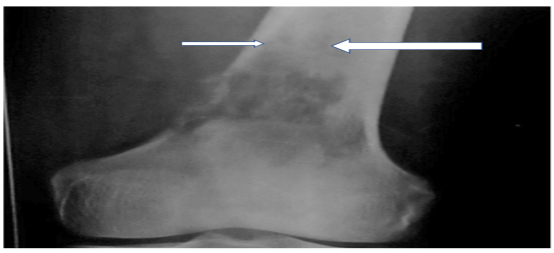
On contrast CT/MR, there is mild to marked enhancement is observed. The enhancement is seen as thin peripheral and or septal (less than2mm), thick peripheral and or septal (>2mm) without nodularity. Peripheral nodular, central nodular and diffuse pattern may be seen. Codman's triangle, malignant periosteal reaction may also be seen [Fig-6]. PR is observed in all cases. Pathological fracture is seen in 25-30%. We had no case with pathological fracture.

FIG 6 18M TOS IN DISTAL FEMUR.SAGITTAL,AXIAL STIR IMAGES SHOWING LAMILLATED PERIOSTEAL REACTION (ARROW)



Another feature is oblique parallel striations in the shafts across the cortical bone in oblique direction due to hypertrophied veins of cortex[6] [Fig-7].

FIG 7 13M PARALLEL STRIATIONS DUE TO HYPERTROPHIED VEINS DISTAL TO GEOGRAPHIC LYSIS



TOS mimics ABC and was known as bone aneurysm. 90% of tumor consists of haemorrhage /necrotic cavities appearing as ABC. The distinct features of TOS are aneurysmal dilatation and fluid-fluid level (FFL). There are certain features which differentiate TO S from ABC.

Table-3: Differences Between ABC And TOC

FEATURES	ABC	TOS
Expansion	Marked	Marked
Zone of transition	Narrow, sclerosis present	Usually wide
Soft-tissue component	Diffuse, well defined with a capsule	Usually focal, ill defined
FFL	Large and occupy most of tumor	Small, non-uniform and occupy less portion of tumor
Septae	Thin smooth	Thick, nodular
Osseous, calcific matrix	Absent	Present

Response to chemotherapy is marked by reduction in size of tumor, reduction in inflammatory response and reduced vascularity.

CONCLUSION

Telangiectatic osteosarcoma is rare variety of osteosarcomas of the extremity with an incidence of 17%. Since it is a great mimicker of ABC from which it is to be differentiated. Lytic lesion, wide zone of transition with malignant periosteal reaction is usually observed in TOS. FFLs are usually small, variable size, irregular septae. Sclerosis is difficult to appreciate but when seen it is diagnostic of TOS.

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